Childhood Nephrotic Syndrome

Childhood nephrotic syndrome can occur at any age but is most common between the ages of 1½ and 5 years. It seems to affect boys more often than girls.

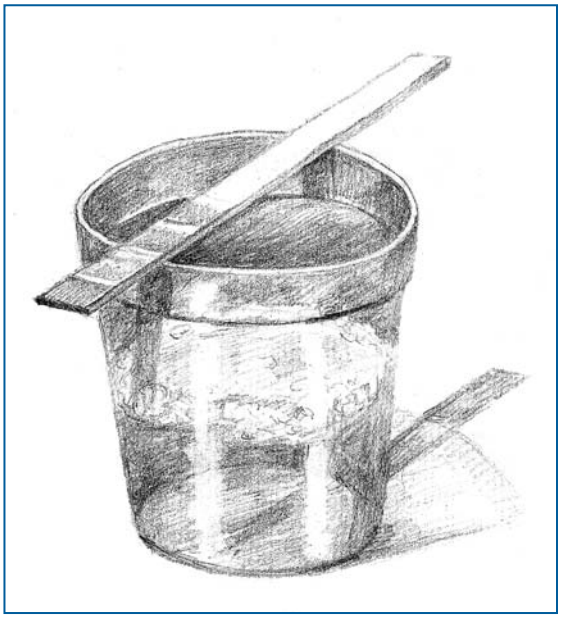
A child with the nephrotic syndrome has these signs:

- high levels of protein in the urine
- low levels of protein in the blood
- swelling resulting from buildup of salt and water

The nephrotic syndrome is not itself a disease. But it can be the first sign of a disease that damages the kidney's tiny blood-filtering units, called glomeruli, where urine is made.

The kidneys are two bean-shaped organs found in the lower back. They are about the size of a fist. They clean the blood by filtering out excess water and salt and waste products from food. Healthy kidneys keep protein in the blood, which helps the blood soak up water from tissues. But kidneys with damaged filters may let protein leak into the urine. As a result, not enough protein is left in the blood to soak up the water. The water then moves from the blood into body tissues and causes swelling. You may see swelling around your child's eyes, belly, and legs. Your child may urinate less often than usual and may gain weight from the excess water.

To diagnose childhood nephrotic syndrome, the doctor may ask for a urine sample to check for protein. The doctor will dip a strip of chemically treated paper into the urine sample. Too much protein in the urine will make the paper change color. Or the doctor may ask for a 24-hour collection of urine for a more precise measurement of the protein and other substances in the urine.



A strip of chemically treated paper will change color when dipped in urine with too much protein.

National Institute of Diabetes and Digestive and Kidney Diseases
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The doctor may take a blood sample to see how well the kidneys are removing wastes. Healthy kidneys remove creatinine and urea nitrogen from the blood. If the blood contains high levels of these waste products, some kidney damage may have already occurred. But most children with the nephrotic syndrome do not have permanent kidney damage.

In some cases, the doctor may want to examine a small piece of the child's kidney under a microscope to see if something specific is causing the syndrome. The procedure of collecting a small tissue sample from the kidney is called a *biopsy,* and it is usually performed with a long needle passed through the skin. The child will be awake during the procedure and receive calming drugs and a local painkiller at the site of the needle entry. A patient who is prone to bleeding problems may require open surgery for the biopsy. General anesthesia will be used if surgery is required. For any biopsy procedure, the child will stay overnight in the hospital to rest and allow the health care team to address quickly any problems that might occur.

Minimal Change Disease

The most common form of the nephrotic syndrome in children is called *minimal change disease*. Doctors do not know what causes it. The condition is called minimal change disease because children with this form of the nephrotic syndrome have normal or nearly normal biopsies. If your child is diagnosed with minimal change disease, the doctor will probably prescribe prednisone, which belongs to a class of drugs called corticosteroids. Prednisone stops the movement of protein from the blood into the urine, but it does have side effects that the doctor will explain. Following the doctor's directions exactly is essential to protect your child's health. The doctor may also prescribe another type of drug called a diuretic, which reduces the swelling by helping the child urinate.

When protein is no longer present in the urine, the doctor will begin to reduce the dosage of prednisone. This process takes several weeks. Some children never get sick again, but most do develop swelling and protein in the urine again, usually following a viral illness. However, as long as the child continues to respond to prednisone and the urine becomes protein free, he or she has an excellent long-term outlook without kidney damage.

Children who relapse frequently, or who seem to be dependent on prednisone or have side effects from it, may be given a second type of drug called a cytotoxic agent. The agents most frequently used are cyclophosphamide and chlorambucil. After reducing protein in the urine with prednisone, the doctor may prescribe the cytotoxic agent for a while. Treatment with cyclophosphamide and chlorambucil usually lasts for 8 to 12 weeks. Alternatively, cyclosporine, a drug also used in transplant patients, may be given. Treatment with cyclosporine frequently continues over an extended period.

In recent years, doctors have explored the use of mycophenolate mofetil (MMF) instead of cytotoxic agents for children who relapse frequently. MMF is an immunosuppressant used to treat autoimmune diseases and to keep the body from rejecting a transplanted organ. MMF has not been tested for treating nephrotic syndrome in large clinical trials, but doctors report promising results with small numbers of patients. MMF has milder side effects than cytotoxic agents, but taking immunosuppressants can raise the risk of infection and other diseases. The good news is that most children "outgrow" nephrotic syndrome by their late teens with no permanent damage to their kidneys.

Other Conditions That Involve the Childhood Nephrotic Syndrome

In about 20 percent of children with the nephrotic syndrome, the kidney biopsy reveals scarring or deposits in the glomeruli. The two most common diseases that damage these tiny filtering units are focal segmental glomerulosclerosis (FSGS) and membranoproliferative glomerulonephritis (MPGN).

Since prednisone is less effective in treating these diseases than it is in treating minimal change disease, the doctor may use additional therapies, including cytotoxic agents. Recent experience with a class of drugs called ACE inhibitors, a type of blood pressure drug, indicates that these drugs can help to prevent protein from leaking into the urine and keep the kidneys from being damaged in children with the nephrotic syndrome.

Very rarely, a child may be born with congenital nephropathy, a condition that causes the nephrotic syndrome. The most common form of this condition is congenital nephropathy of the Finnish type (CNF), inherited as an autosomal recessive trait. Another condition that causes nephrotic syndrome in the first months of life is diffuse mesangial sclerosis (DMS). The pattern of inheritance for DMS is not as clearly understood as the pattern for CNF, although the condition does appear to be genetic.

Since medicines have little effect on congenital nephropathy, transplantation is usually required by the second or third year of life, when the child has grown sufficiently to receive a kidney. To keep the child healthy, the doctor may recommend infusions of the protein albumin to make up for the protein lost in urine and prescribe a diuretic to help the child eliminate the extra fluid that causes swelling. The child's immune system may be weakened, so antibiotics should be given at the first sign of infection.

Congenital nephropathy can disturb thyroid activity, so the child may need a substitute hormone, thyroxine, to promote growth and help bones mature. A blood thinner like warfarin may be necessary to keep the child's blood from clotting.

A child with congenital nephropathy may need tube feedings to ensure proper nutrition. In some cases, the diseased kidney may need to be removed to eliminate proteinuria. Dialysis will then be required to replace kidney function until the child's body is big enough to receive a transplanted kidney. Peritoneal dialysis is preferable to hemodialysis for young children.

In peritoneal dialysis, a catheter is surgically placed in the child's abdomen and then used to introduce a solution into the abdominal cavity (the peritoneum). The solution draws wastes and extra fluid from the child's blood stream. After a few hours, the solution is drained and replaced with a fresh supply. The drained solution carries the waste and extra fluid out of the child's body.

Hope Through Research

The National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) conducts and supports research to help many kinds of people with kidney disease, including children. NIDDK's Division of Kidney, Urologic, and Hematologic Diseases (DKUHD) maintains the Pediatric Nephrology Program, which supports research into the causes, treatment, and prevention of kidney diseases in children. In 2002, DKUHD initiated the Focal Segmental Glomerulosclerosis Clinical Trial to learn more about the best way to treat FSGS. Then, in 2003, DKUHD initiated the Prospective Study of Chronic Kidney Disease in Children to learn more about the negative effects of pediatric kidney disease, including cardiovascular disease and neurocognitive impairment.

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This fact sheet is also available at *www.kidney.niddk.nih.gov.*

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